

Dyke-Davidoff-Masson in a pregnant woman: A rare case report

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ABSTRACT

Background: The first examples of this syndrome were identified and described by Dyke, Davidoff and Masson in 1933. The global incidence of the condition is still uncertain, even though 100 cases have been reported worldwide almost a century after its discovery. Clinical symptoms include mental retardation, hemiparesis or hemiplegia, facial or bodily asymmetry with atrophy and localised and/or widespread drug-resistant epilepsy. **Case description:** 23-year-old G2A1 (gravid 2 abortion 1) presented with history of amenorrhoea since 9 months and pain in abdomen since 6 hours and she late developed complex partial status epilepticus and had a history of recurrent seizures since 10 years of age. MRI brain was done which revealed Dyke Davidoff Masson Syndrome. **Conclusion:** DDMS, a condition characterized by frequent and persistent seizures, is rare and can be difficult for medical professionals to diagnose accurately due to its rarity. However, specific imaging findings related to this disease can be revealed using MRI.

Keywords: Dyke Davidoff Masson Syndrome (DDMS), Seizures, Epilepsy, MRI Brain

1. INTRODUCTION

Hemiatrophy, also known as Dyke-Davidoff-Masson syndrome (DDMS), is characterised by hypoplasia or hypoplasia of one cerebral hemisphere and is frequently caused by a lesion to the developing brain during pregnancy or the first few months of life. Depending on the severity of the brain lesion, different clinical traits apply. Some of their most prevalent symptoms are recurrent seizures, contralateral hemiplegia and asymmetry of face, mental impairment or difficulty in learning and speech and language issues. There have been isolated cases of sensory loss and psychological symptoms resembling schizophrenia (Behera et al., 2012).

It was first discovered by Dyke, Davidoff and Masson in nine hemiplegic patients. By seeing alterations in plain skull X-rays, they were able to explain the neurologic aspects (Ayele and Zewde, 2019). There may also be concurrent calvarial thickening and hyper pneumatization of the frontal sinuses, which help to distinguish it from other hemi atrophic illnesses such as Rasmussen's encephalitis and Sturge-Weber syndrome (Acharya et al., 2022).

It is characterized by:

- Strengthening of the cranial vault (compensatory)
- Elevation of the petrous ridge;
- Enlargement of the frontal sinus, as well as ethmoidal and mastoid air-cells
- Ipsilateral displacement of the falx
- Capillary malformations (a novel finding in kids with the Dyke-Davidoff-Masson syndrome) (Ho et al., 2023)

2. CASE PRESENTATION

A 23-year-old G2A1 (gravida 2 abortion 1) with 36+1 came to emergency department of tertiary care centre with complaint of pain in abdomen since 6 hours. She was a known case of epilepsy since 10 years. As per her mother, she was late preterm with triple loop of cord around neck and was in intensive care unit for management of hypoxic insult. She also faced difficulty in achieving milestones due to developmental delays. There was no family history associated with seizures.

She was on Tab Levitriacetam 500 mg BD, Tab Lacosemide 100mg BD, Tab Clobazam 10mg, Tab Carbamazepine 600 mg BD since 10 years. She was also a known case of hypothyroidism and sickle cell pattern. She was previously hospitalised for status epilepticus before pregnancy. She got intubated in view of fall in saturation and was in coma for 4 days. She also had covid-19 positive 2 years ago. CT brain was done (scans not available). Per abdomen examination revealed uterus of 34 weeks size, relaxed, cephalic, fetal heart rate were regular with baseline 160 beats per minute. On per vaginum, os was closed and uneffaced. Neurological examination revealed mild right-sided hemiparesis with a power of 4/5, extensor right plantar reflex and hyperflexia. No facial atrophy noticed. Cranial nerve and sensory examination were unremarkable (Figure 1, 2, 3, 4).

All routine investigations were normal. An ultrasound scan showed a live single intrauterine fetus at about 36-week gestation, with an estimated fetal weight of 2700+/-300g, an adequate amniotic fluid measuring 10cm and a normal Doppler. She was admitted in high-risk obstetrics ward and daily non stress test was done for fetal well-being. She had 1 episode of seizure in hospital which was sudden in onset and was associated with a confused state and motor automatisms, frothing from mouth and twitching of leg. She was managed by Levitriacetam 1 gram injection, Injection Lacosemide 200mg BD, Tab Clobazam 10mg, Tab carbamazepine 600 mg BD. At 36+6 weeks of gestational age her emergency caesarean section was taken in view of persistent fetal tachycardia. In presence of multispeciality team section was performed. Intraoperatively everything was normal and female of 2.5kg was extracted which later shifted to mother side. On day 6 of post-natal care MRI Brain was done which revealed:

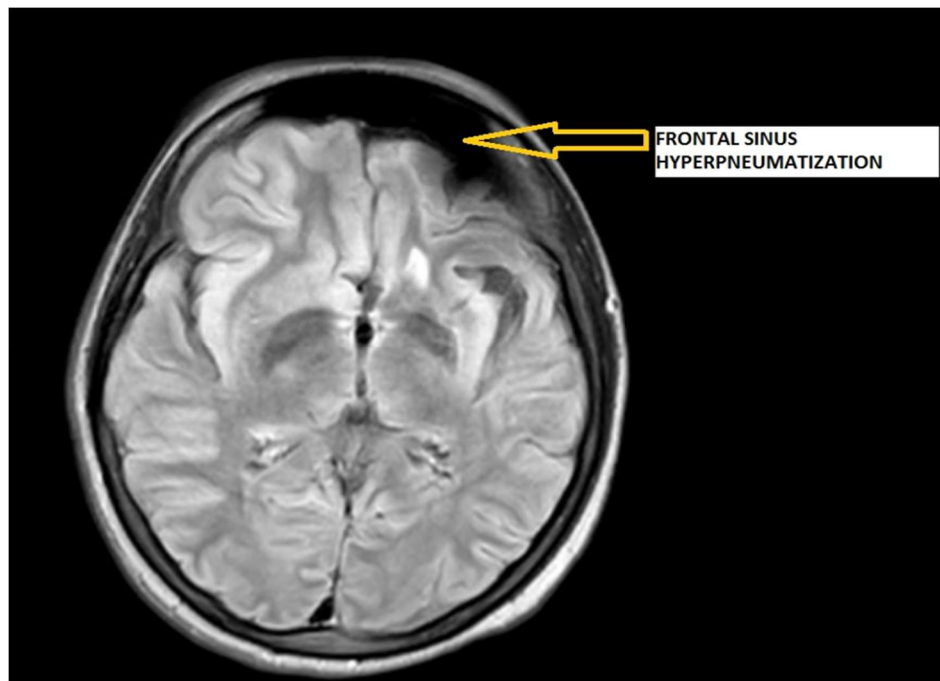


Figure 1 MRI of Brain showing Flair image of Frontal sinus Hyperpneumatization

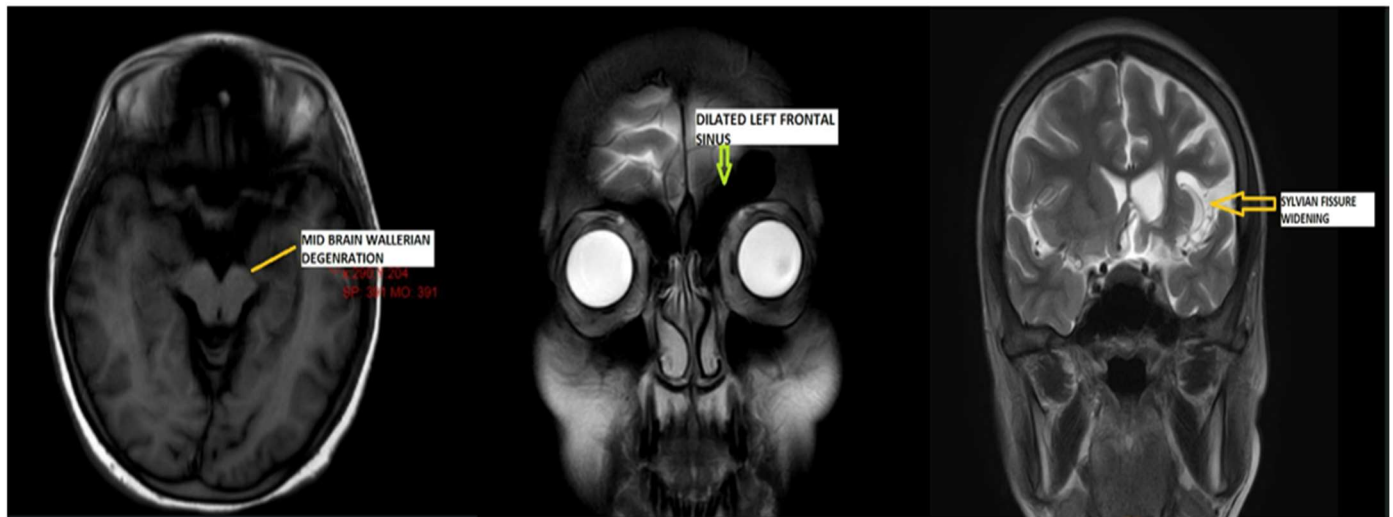


Figure 2 MRI of Brain showing T2 image of Mid Brain Wallerian Degeneration, Dilated left Frontal Sinus and Sylvian fissure Widening

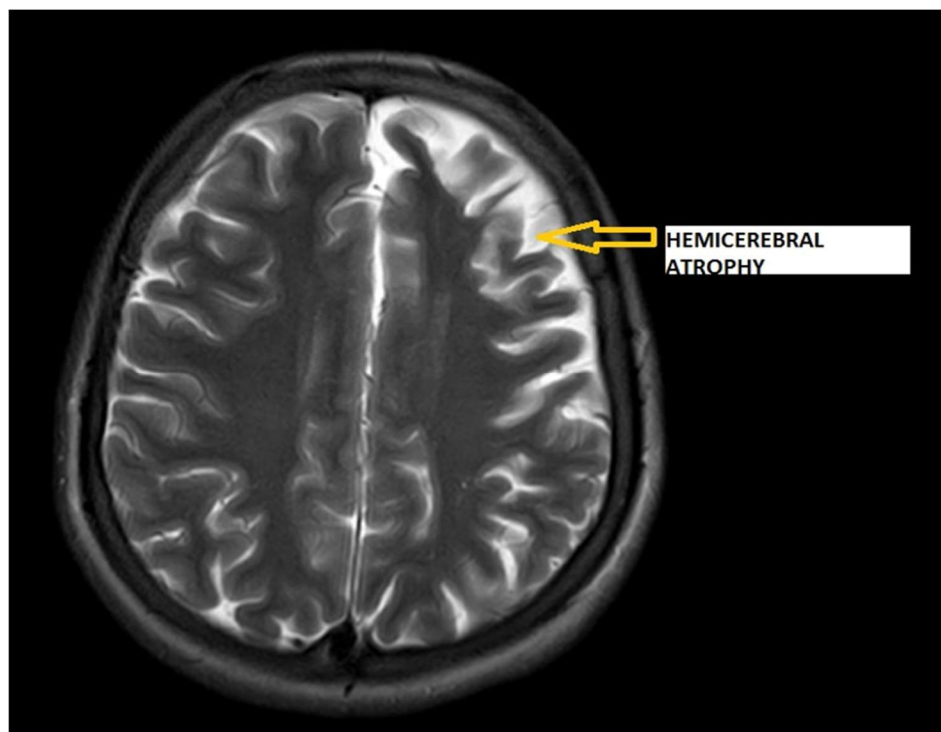


Figure 3 MRI of Brain showing T2 image of Hemicerebral Atrophy

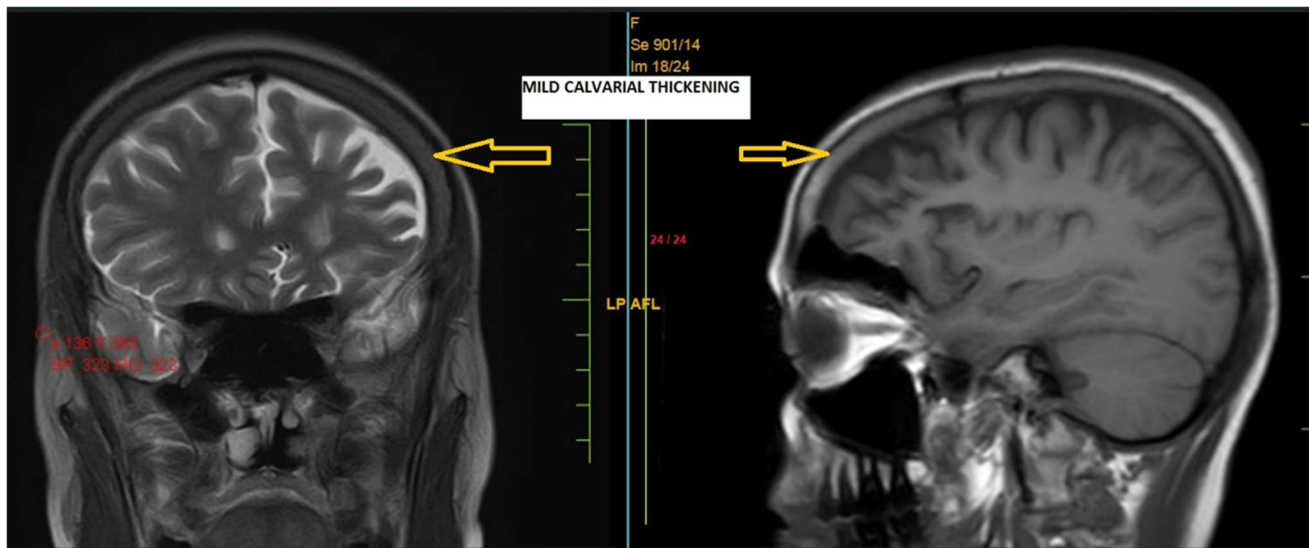


Figure 4 MRI of Brain showing Coronal T2 and sagittal T1 image of mild Calvarial Thickening

3. DISCUSSION

The first case of this syndrome was identified and described by Dyke, Davidoff and Masson in 1933 (Ho et al., 2023). Nearly a century later, there have been 100 cases reported globally, but the overall incidence is still unclear. The International League against Epilepsy classifies DDMS as epilepsy with anomalies of the brain's structural makeup (ILAE). The clinical symptoms include mental retardation, hemiparesis or hemiplegia, facial or body asymmetry with atrophy and focal and/or widespread drug-resistant epilepsy. In addition to these symptoms, uncommon individuals may also present with cerebellar atrophy, neuropsychiatric conditions and ear malformations (Ho et al., 2023). Both sexes and any hemisphere may be impacted, however left side involvement and male gender are more prevalent (Behera et al., 2012).

Our patient is a 23-year-old gravida 2 abortion 1 unbooked with 36+1 weeks of gestational age who initially presented with pain in abdomen and next day she developed headache, followed by episodic complex partial seizures. Due to persistent fetal tachycardia caesarean section done. On day 6 of PNC, she had recurrent episodes of seizures for which MRI Brain was done which revealed DDMS. Later she was kept on same anticonvulsant drugs but dose of each drug was increased.

A thorough clinical examination, an accurate history and the results of the radiology are used to make the correct diagnosis. This condition needs to be separated from Fishman syndrome, Rasmussen encephalitis, Sturge Weber syndrome, Silver-Russel syndrome, linear nevus syndrome, basal ganglia germinoma and more. In addition to the clinical presentation, the radiological outcomes of CT scans and MRIs are essential in making the diagnosis and assessing the severity of the illness. These outcomes can vary from patient to patient, although they typically involve atrophy of the cerebral hemisphere adjacent to the lesion, conspicuous sulci and lateral ventricular enlargement (Behera et al., 2012).

Patients with DDMS typically have refractory seizures when they are first diagnosed, thus the focus of treatment should be on controlling the seizures with the right anticonvulsants. There are situations when several anticonvulsants are used. In addition to medication, speech, occupational and physical therapy are important components of the children long-term care. The prognosis is better if hemiparesis develops after the age of two and without a prolonged or recurrent seizure. With an 85% success rate in some cases Acharya et al., (2022), hemispherectomy is the therapy of choice for kids with incapacitating debilitating seizures and hemiplegia.

4. CONCLUSION

DDMS is a rare condition that causes frequent, on-going seizures. Medical professionals could easily misdiagnose it because it is a rare disease. MRI can be used to show specific imaging findings related to this disease. This is the first instance of a pregnant woman having DDMS. To suspect and diagnose DDMS, one must have a thorough understanding of the clinical presentation, related risk factors, such as birth asphyxia, cerebral haemorrhage, infections and imaging findings.

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Informed consent

Not applicable.

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Conflict of interest

The authors declare that there is no conflict of interests.

Data and materials availability

All data sets collected during this study are available upon reasonable request from the corresponding author.

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